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Successful Management of Neutropenia in a Patient with CD40 Ligand Deficiency by Immunoglobulin Replacement Therapy

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Abstract:

Hyper-IgM syndromes are characterized by profound reduction of serum IgG, IgA, and IgE levels with normal or increased concentrations of serum IgM. CD40 ligand deficiency is X-linked form of the disease, which results in a lack of immunoglobulin class switching from IgM to IgG in B cells. In addition to the recurrent infections, a number of patients suffer from neutropenia. There are some evidences indicating the effect of G-CSF in combination with intravenous immunoglobulin (IVIG) in improvement of neutrophil counts, which has become the most common procedure to control neutropenia.

In this report we present a 6 year-old patient of CD40 ligand deficiency, who suffered from chronic, severe neutropenia. Administration of IVIG was started for him when the diagnosis was made at the age of 1.5 years and he was on the regular IVIG therapy after that time untill now for a period of 4.5 years. IVIG and prophylactic antibiotic therapy, despite cessation of granulocyte colony-stimulating factor, injection after one month, corrected the severe neutropenic state of this patient.

It seems that regular administration of sufficient doses of IVIG can be useful in the management of neutropenia in CD40 ligand deficiency, which results in better quality of life with decreasing occurrence of infection.

Keywords:

CD40 Ligand ، Hyper IgM Syndrome

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